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An unusual case of deafness with speech impairment: lesson in diagnosis and management

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Abstract
Hearing impairment can be the cause of significant disability and handicap. This medico-legal case demonstrates the need for accurate assessment of both the severity and type of hearing loss if the best clinical management is to be provided. In particular, the case identifies the critical role of additional, objective auditory testing when pure tone audiometry, which depends on the subjective response of the individual, is inconsistent or indicates severe hearing impairment.

Key words: Hearing loss, functional; Audiometry, evoked response.

Case report

A 48-year-old woman was seen by her family doctor on several occasions over a five-week period, complaining of deafness, tinnitus and earache in both ears. She was under psychiatric supervision for a personality disorder, and her family doctor considered that her symptoms were likely to have an hysterical origin. When her symptoms persisted however she was referred to the ENT Department: an audiogram in April 1991 (Table I) showed a mild hearing loss bilaterally (but was reported as being 'unreliable') and her earache diagnosed as being of temporo-mandibular joint origin. She was referred to an oral surgeon and given a two-month follow-up appointment to retest her hearing.

Meanwhile she was reviewed by her psychiatrist and an assessment period at the local psychiatric day hospital arranged. On this occasion, it was noted that she appeared not to hear, that her speech had altered, and arrangements were made by the patient to enrol at the local Deaf Centre to learn sign language.

She sought a further ENT opinion, this time describing a sudden, total loss of hearing on the left and a less severe, progressive loss on the right. A past history of head injury, 10 years previously, was elicited. When audiometry showed a deterioration of hearing thresholds on the right and a total loss of hearing on the left in June 1991 (Table I) she was admitted for further investigation and bed-rest with a presumptive diagnosis of perilymph fistula. A week later, following repeat audiometry (Table I) she underwent an exploratory myringotomy of the right ear following which a possible perforation of the round window membrane was patched, although no definite diagnosis of a perilymph leak could be made.

This produced no clear-cut benefit and six weeks later in August 1991 she represented with a total loss of hearing bilaterally (Table I). An infective/inflammatory aetiology was now considered and she was treated with antibiotics and steroids. This resulted in only a limited improvement (Table I) and the patient was referred for cochlear implant assessment. At this point she declined further clinical or operative intervention and one year later she initiated a medico-legal claim against her former GP and her District Health Authority for failing to detect and advise her in connection with hearing difficulties in consequence of which she became deaf. Note that the sequential audiometry (Table I) had been performed by different technicians within the same Department of Audiology.

Medico-legal interview

At the medico-legal interview in February 1993 (18 months after initiating the claim and at a different centre), the patient gave the impression clinically of being profoundly deaf. She was wearing a vibrotactile aid on her wrist, attempting to lip read, and appeared to require gestures from her partner in order to understand the questions asked. Her speech was strikingly abnormal, superficially consistent with an acquired profound hearing loss. When tested 'behaviourally' by clinking a teaspoon in

TABLE I
SEQUENTIAL AUDIOMETRY PRIOR TO MEDICO-LEGAL ASSESSMENT

Date	250 Hz	500 Hz	1 kHz	2 kHz	4 kHz	8 kHz
Right ear dBHL						
08/04/91		20	25	25	5	
10/06/91	90	85	70	80	85	100
17/06/91	105	100	80	95	100	
06/08/91	NR	NR	NR	NR	NR	NR
28/11/91	50	90	115	110	105	90
Left ear dBHL						
08/04/91		45	35	25	10	
10/06/91	NR	NR	NR	NR	NR	NR
17/06/91	NR	NR	NR	NR	NR	NR
06/08/91	NR	NR	NR	NR	NR	NR
28/11/91	110	120	110	110	NR	NR

NR = No response.

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TABLE II
MEDICO-LEGAL ASSESSMENT: PURE TONE AUDIOMETRY (PTA), ACOUSTIC REFLEX THRESHOLDS AND CORTICAL EVOKED POTENTIAL THRESHOLDS

15/02/93	250 Hz	500 Hz	1 kHz	2 kHz	4 kHz	8 kHz
Right ear dBHL						
PTA	VTR	VTR	NR	NR	NR	NR
ART ipsi		95	90	95		
ART contra		100	95	95	85	
CEPT			10		15	
Left ear dBHL						
PTA	VTR	VTR	NR	NR	NR	NR
ART ipsi		90	85	90		
ART contra		95	90	90	90	
CEPT			10		10	

VTR = Vibrotactile responses.
ART = Acoustic reflex thresholds.
CEPT = Cortical evoked potential thresholds.
NR = No response.

a coffee cup immediately behind her head, she failed to turn. However, to confuse this clinical impression, she was wearing an ear plug in the right ear 'to stop the noise'.

The history elicited was essentially the same as previously, but in addition she gave further details of her psychosocial background, which included being battered, first by her father (with whom she was thought to have had an incestuous relationship) and then by her husband.

Audiological assessment

Her otological examination in February 1993 was normal, and she was investigated using a battery of auditory tests to establish, if possible, the severity and type of her hearing loss.

Standard pure tone audiometry (PTA) performed immediately following the medico-legal interview, revealed absent hearing thresholds (Table II) with vibrotactile responses only at lower frequencies. Impedance testing showed normal middle ear pressures with normal compliance of the tympanic membranes bilaterally, and normal acoustic reflex thresholds both ipsilaterally and contralaterally (Table II).

Brain stem auditory evoked potentials were normal bilaterally at 90 dBHL stimulus intensity levels and the central auditory conduction time between waves I and V was within normal limits (Figure 1). Cortical auditory evoked potentials (recorded and analysed blind to the audiometric evaluation) were also normal and could be traced down to thresholds of 10 and 15 dB at 1 and 4 kHz respectively on the right, and to 10 dB at both 1 and 4 kHz on the left (Figure 2a, b, c and d).

Otoacoustic emissions (recorded using the otodynamics ILO 88 analyser) were normal at 5.3 dB on the right and at 11.1 dB on the left (Figure 3).

Speech assessment

An assessment of the patient's speech was made retrospectively by one of the authors (A.O.) from an interview recorded on video.

The patient presented difficulties in both the reception and production of speech, which were striking for their inconsistencies. Her speech production contained several features, both segmental (i.e. relating to discrete speech sounds such as vowels or consonants) and suprasegmental (i.e. features of speech longer than a segment such as rhythm or voice quality) which were typical of speech deterioration resulting from acquired hearing loss (Cowie and Douglas-Cowie, 1992). Her segmental speech contained examples of deterioration of alveolar sounds, typically affected by acquired hearing loss, but showed inconsistencies: the same sound was produced both correctly and incorrectly i.e. the word 'said' was produced correctly but also as 'taid', 'stairs' and 'stared'. Her suprasegmental speech demonstrated typical changes in pitch and intonation, but features such as loudness control, voice quality and rhythm, remained intact.

The speech pattern was unusual for the discrepancy between the degree of segmental and suprasegmental deterioration. At the segmental level there was severe deterioration i.e. 's' was said as 'd' and thus a voiceless fricative had become a voiced plosive, a change in both voice and manner of articulation, whereas the suprasegmental changes were mild.

Discussion

The medical negligence claim was based on the assumption that the diagnosis of profound hearing loss was correct and that negligence lay in failing to detect it earlier and advise her appropriately. Despite her clinical appearance, it is clear from the results of the auditory tests carried out at her medico-legal assessment, that she had normal auditory function and that her failure to hear was not as a result of cochlear damage as originally assumed.

This error appeared to arise from the belief that thresholds obtained from pure tone audiometry gave an accurate indication of her hearing status, and that there was no reason to suspect that she did not have a profound hearing loss given her clinical presentation and markedly abnormal speech. However, her psychiatric history should have aroused suspicion and further objective evidence of hearing impairment should have been sought. Reliance on the results of pure tone audiometry led to an unfortunate and inappropriate path of clinical management.

The additional auditory tests performed on this patient at medico-legal assessment i.e. measuring the acoustic reflex thresholds, the mechanical and the electrical

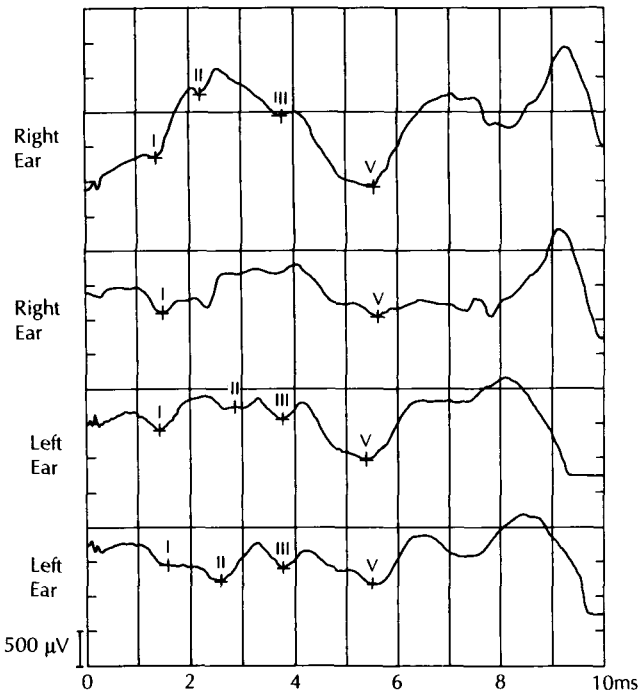

FIG. 1
Auditory brain stem evoked potentials recorded at 90 dB during medico-legal assessment.

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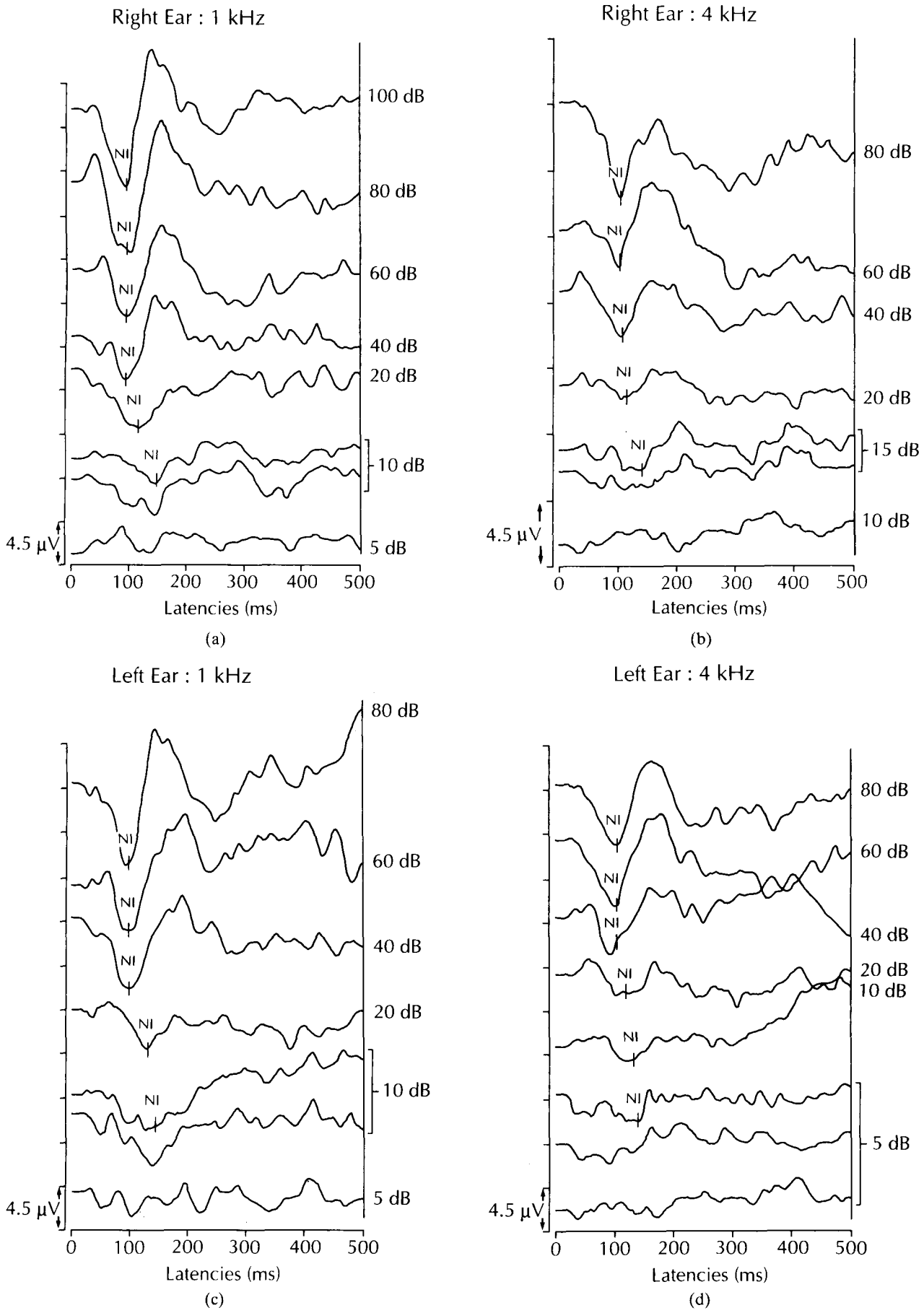

FIG. 2 (a, b, c, and d)
Cortical evoked potentials at medico-legal assessment.

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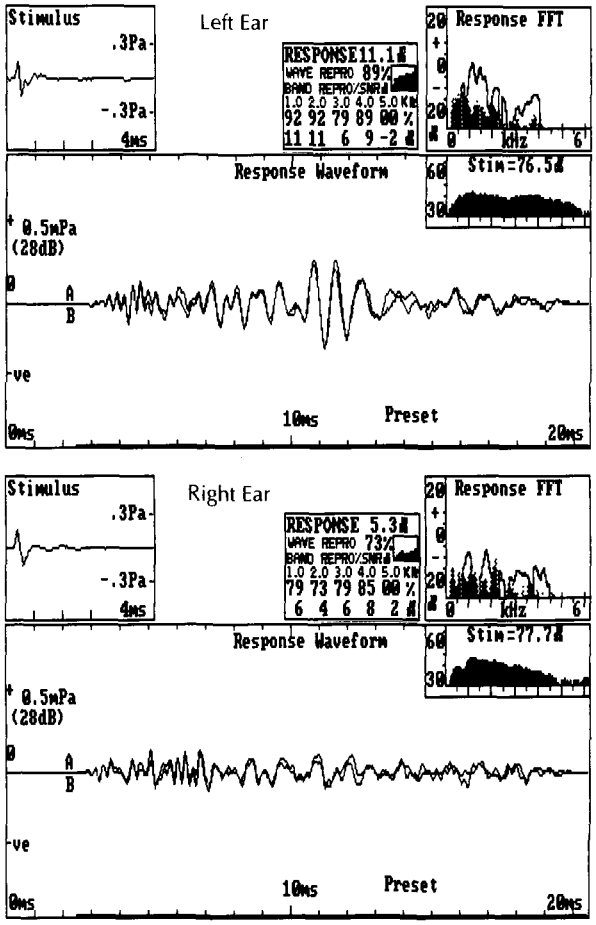

FIG. 3

Otoacoustic emissions at medico-legal assessment.

responses of the auditory system, gave objective evidence of normal auditory function at different sites in the auditory pathways.

The majority of these tests should be routinely available in good audiology departments. Recording of otoacoustic emissions would suggest normal functioning of the outer hair cells of the cochlea and imply hearing thresholds of 40 dB or better. Note that this technology has been embraced by the National Institutes of Health as the method of choice for screening neonates for hearing loss (Collet *et al.*, 1990). Measurement of stapedius reflex thresholds (Prasher and Cohen, 1993) during impedance testing demonstrates the integrity of the VIII/VIIth nerve reflex arc and immediately indicates that hearing thresholds must be at least as good as the values recorded. Measurement of normal brain stem auditory evoked potentials with normal wave I and V latencies at 90 dB indicates thresholds at 2–4 kHz of at least 90 dB or better (Prasher and Gibson, 1993).

Less widely used (although the technology is available in most departments) is the measurement of cortical auditory evoked potentials, which although more time-consuming have greater significance medico-legally as they are the most accurate objective measure of frequency-specific hearing thresholds and give the closest approximation to the true pure tone audiogram (Prasher *et al.*, 1993). The normal results in this patient provide strong evidence for the integrity of auditory pathways from the middle ear rostrally through to the auditory cortex bilaterally, and suggest that the hearing loss recorded during pure tone audiometry was 'functional' in nature.

The altered speech of this lady, with characteristics of a severely hearing-impaired patient was very misleading. The acquisition of such a speech pattern implies either a deliberate and sophisticated attempt to conceal the truth about her hearing or a hysterical conversion phenomenon with subconscious denial of auditory input.

Although this case is presented with the advantage of hindsight and the privilege of the expert witness of access to GP and hospital medical records (which documented a psychiatric admission 20 years previously for aphonia and mutism), the underlying issue of good clinical practice remains. When, as in this case, both clinical assessment and subjective testing with pure tone audiometry suggests a severe to profound hearing loss, or when hearing thresholds are reported as unreliable, more sophisticated objective tests are required to confirm the hearing loss to avoid the sort of pitfalls in clinical management described above.

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